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# Simplified surgical-hybrid Melody® valve implantation for paediatric mitral valve disease

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## Abstract

Children suffering from left atrioventricular valve (LAVV) disease not amenable to repair represent a significant challenge. The results of surgical reconstruction are not optimal. Valve replacement as an alternative is associated with poor results. The surgical-hybrid approach with implantation of a stented biological valve (bovine jugular vein graft, Melody® valve) seems to represent a new therapeutic option. Here we demonstrate our case, the consideration and the approach to extreme clinical findings in a small child. We describe a simplified surgical-hybrid Melody valve implantation in a LAVV position. The technique of implantation is relatively simple and the immediate post-operative result very good.

**Keywords:** Congenital mitral valve stenosis • Mitral valve replacement • Melody® valve implantation • Hybrid approach

## INTRODUCTION

Children suffering from left atrioventricular valve (LAVV) disease represent a significant challenge. Surgical implantation of a Melody® (Medtronic, Minneapolis, MN, USA) valve in the mitral position recently described by Abdullah *et al.* [1] and Quiñonez *et al.* [2] appears to be a promising option.

We describe a simplified surgical/hybrid implantation of a Melody® valve in the mitral position in a small child suffering from stenosis, 2 years after repair of a complete AV canal defect.

## CASE REPORT

A 2.5-year-old boy was admitted to our hospital with a severe LAVV stenosis nearly 2 years after a satisfactory correction of a complete AV canal which coincided with the diagnosis of acute myeloid leukaemia.

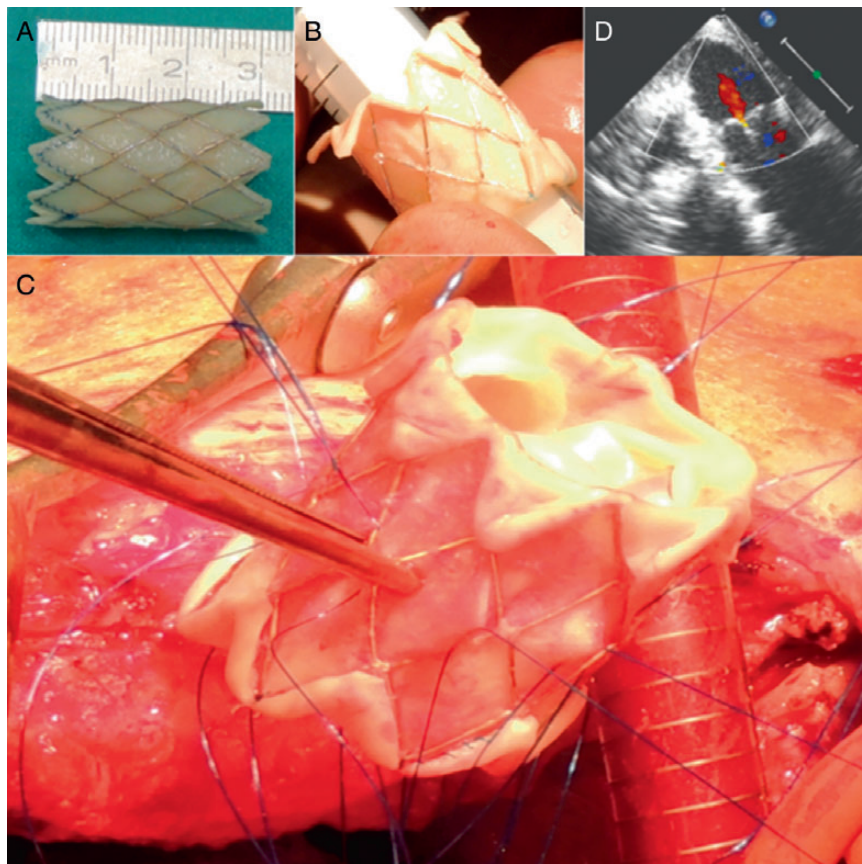
The aetiology of the stenosis was multifactorial and attributed to: hypoplastic annulus, skewed commissures, thickened anterior leaflet and extreme subvalvular crowding. The need for the relief of stenosis became more urgent since chemotherapy for acute myeloid leukaemia was expected to lead to significant volume overload.

Percutaneous balloon valvuloplasty resulted only in transient improvement. A surgical repair was performed, which included extensive commissurotomy, papillary muscle and chordal splitting, partial resection of the redundant thickened leaflet as well as lengthening of the anterior leaflet with autologous pericardium. Intraoperatively, the annulus measured around 12 mm (z-score: -2.3).

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The little improvement after surgical reconstruction was short-lived with the mean gradient quickly rising to the preoperative level.

A second reoperation was performed after 3 weeks. The LAVV was accessed trans-septally. The incision across the anterosseptal tricuspid valve commissure was extended across the ventricular septal defect patch. Replacement with a Melody® valve was undertaken. The original 3-cm long Melody® valve was shortened to ~2 cm by bending the proximal and distal rows of prongs (Fig. 1A and B). The Melody® valve was crimped (12 mm diameter) and annular fixation was achieved using 10 single 5-0 polypropylene stitches, most of them hooking the wired stent (Fig. 1C). Simple interrupted sutures were placed to facilitate a later dilatation. Part of the subvalvular apparatus abutting the anterior commissure was detached, while the complete anterior leaflet and the corresponding subvalvular apparatus were retained. With this technique, equal protrusion of the Melody® valve into the left atrium and ventricle was ensured. Further stabilization was achieved by progressive dilatation using a 14- and 16-mm balloon. Therefore, any kind of further anchorage of the stented valve (pericardial sewing cuff or a distal fixation to the left ventricular lateral wall), as described previously [1, 2], was not necessary. The trans-septal opening was reconstructed using a generous-sized xenopericardial patch to enlarge the left atrium and to ensure an unobstructed inflow. Intraoperative echocardiography showed a good result without paravalvular leakage, with a minimal central jet (Fig. 1D). In our first case, the clamping time was 150 min. This was due to the revision of one fixation suture, which had affected the valve motion. Avoiding this and considering that this was our first case, the operation time will be significantly shorter in the future.



**Figure 1:** (A) Melody® transcatheter pulmonary valve. (B) Preparation of the Melody® valve. (C) Fixation of the Melody® valve. (D) Intraoperative transoesophageal echocardiography.

There was no obstruction to the pulmonary venous flow. The invasive blood pressure measurements of the left ventricle and the aorta revealed a gradient of <10 mmHg. Postoperative trans-thoracic echo did demonstrate gradients up to 40 mmHg, which reduced on swapping adrenaline with noradrenaline (reducing the hypercontractility and increasing afterload). In the subsequent course during the first postoperative week, the left ventricular outflow tract gradient stabilized at 10 mmHg. The mean gradient across the Melody® valve remained <5 mmHg and the previous pulmonary venous congestion resolved completely.

Therapeutic heparinization was instituted. Because of spontaneous thrombocytopenia related to the myeloid leukaemia, use of an antiplatelet agent was avoided. Chemotherapy could be initialized 3 weeks after valve replacement without any haemodynamic impairment. Repeated echocardiographic assessments always revealed low Doppler gradients without any relevant regurgitation.

## DISCUSSION

The surgical treatment of mitral valve stenosis in paediatric patients remains very challenging. Understanding the mechanism is the key to a successful reconstruction. A hypoplastic annulus does not have dependable solutions. While the surgical options are limited, balloon valvuloplasty is also palliative. Although improvements may be achieved acutely, there is a high risk of restenosis or severe mitral regurgitation [3].

Since the chances of successful reconstruction or balloon valvuloplasty are uncertain, valve replacement remains the ultima ratio. There exist, however, few options for valve replacement in small children [4]. Mechanical valves are very durable, but need lifelong anticoagulation. Biological valve substitutes such as homograft or xenograft are difficult to get in small sizes (14 or 16 mm), do not grow and degenerate/calcify quickly.

Percutaneous pulmonary valve replacement using stented valves like the Melody® valve or the Edwards SAPIEN® pulmonic valve (Edwards Lifesciences, LLC, Irvine CA, USA) has become an established alternative to surgery [5]. Hence, it seems logical that a stented valve might also be placed in the mitral position, even in children.

The Melody® valve offers many advantages: the circular stent provides a large effective orifice area and hopefully protects from pannus ingrowth. Additionally, the bovine jugular vein leaflet tissue is thin and pliable, which is suitable for the low cardiac output in infants. And most importantly, it could be redilatated corresponding to the growth of the child via a minimally invasive approach.

Our technique is a step forwards in standardizing and simplifying the Melody® valve implantation procedure in a hybrid-surgical setting. In contrast to the reported Boston group approach, our technique avoided resection of the subvalvular left ventricular apparatus which helped to position and stabilize the valve into the correct inflow position, also making a distal fixation dispensable. Furthermore, we did not use an additional pericardial sewing ring to prevent paravalvular leaks. We experienced that

the remaining valvular tissue and the intraoperative redilatation ensured a complete expansion and a leak-proof circular anchoring without paravalvular leakage.

We avoided any resection of stent struts which may influence the radial and longitudinal stability. The bending of the proximal and distal rows of the struts reduces the valve profile and prevents atrial inflow and LVOT obstruction.

While hybrid-surgical implantation of a Melody® valve in the mitral position is an elegant option that can be safely performed and reproduced, the till date published experiences remain anecdotal. In view of this, the indications, technique, impact on pulmonary venous flow, impingement on the LVOT, anticoagulation regimen and long-term durability of the Melody® valve in systemic circulation are questions which await answers pending greater experience.

**Conflict of interest:** Oliver Kretschmar serves as a Melody proctor.

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